Case Report ISSN 2639-944X

Journal of Medical - Clinical Research & Reviews

Bullous Pemphigoid in a Native American/ American Indian Patient

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Received: 29 Nov 2022; **Accepted:** 19 Dec 2022; **Published:** 24 Dec 2022

Citation: Bannon M, Bastible ST, Gaskins T, Rotton DB, Afif M, Kaushik P. Bullous Pemphigoid in a Native American/ American Indian Patient. J Med - Clin Res & Rev. 2022; 6(12): 1-3.

ABSTRACT

Globally, bullous pemphigoid affects less than fifteen people per million per year worldwide. This autoimmune disease occurs most frequently amongst those of northern European ancestry and, most predominantly, in the elderly over sixty-five. While bullous pemphigoid can present as a paraneoplastic syndrome, it can occasionally occur alone. Bullous pemphigoid typically presents with several large blister-like lesions on the skin of flexor surfaces and the trunk. These generally vary in size and stage and are formed after an autoimmune attack on the hemidesmosomes, causing a separation between the basement membrane and the basement membrane cells. Here we present, to our knowledge, the first known case of non-paraneoplastic bullous pemphigoid in an elderly Native American / American Indian patient.

Keywords

Bullous Pemphigoid, Native American / American Indian, BP180 antibodies.

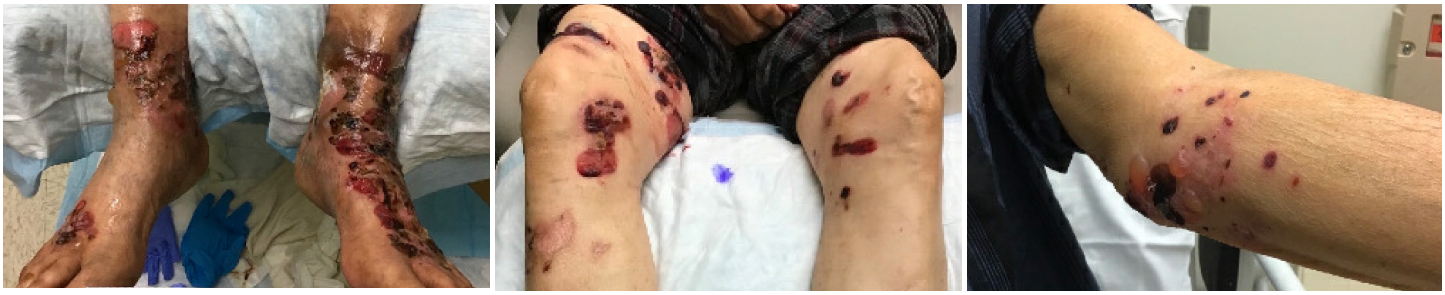
Case Report

An 82-year-old Native American male presented to a surgical wound care clinic with an explosive onset of bullous skin lesions in Figures 1, 2 and 3 below. He had a non-contributory past medical history and no prior history of cancers or smoking, though he used smokeless tobacco for 30 years. A biopsy was taken from the right medial thigh and sent for pathology. He was subsequently referred to a Rheumatologist and Internal Medicine Resident clinic for further evaluation, including a BP180 antibody lab test. Results of the biopsy later returned as antecedent interface dermatitis without the commitment of bullous pemphigoid from the pathologist even on direct immunofluorescence. However, lab results from the BP180 antibody test were elevated, suggesting bullous pemphigoid. Given the high clinical suspicion of bullous pemphigoid in conjunction with the positive antibody and the inconclusive biopsy, the patient was started on the treatment of bullous pemphigoid. He was subsequently initiated on systemic prednisone, and workup for neoplasms was aggressively perused given the association as a paraneoplastic syndrome. After a thoroughly negative cancer screening, doxycycline and methotrexate were added to the patient's therapeutic regimen.

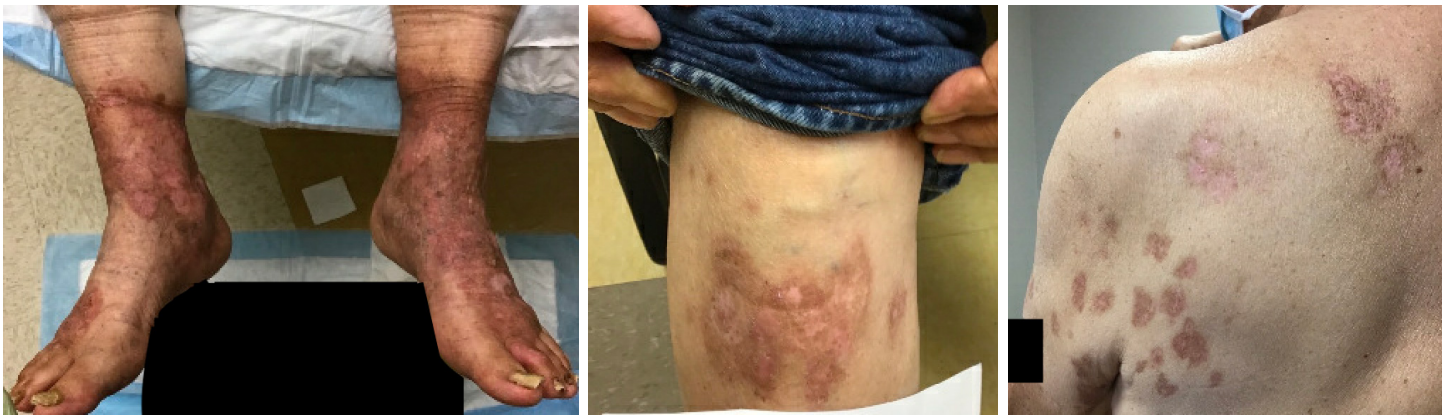
Initially, the patient responded quite well to prednisone 40 mg daily with near complete resolution of his bullae at his follow-up thirty days later. Subsequent addition of disease-modifying antirheumatic drugs (DMARDs) doxycycline and methotrexate resulted in further improvement in symptoms. This combination ultimately stopped the progression of this disease and allowed the patient's systemic steroids to be discontinued. At the time of this case report, the patient had remained bullae-free. Post-treatment photos are shown in figures 4, 5, and 6.

Discussion

Bullous pemphigoid occurs from the autoimmune degradation of



Figures 1-3: Active disease prior to treatment.



Figures 4-6: In remission after treatment

hemidesmosomes. These hemidesmosomes connect the basement membrane cells to the basement membrane. Disruption of this junction is the most common subepidermal autoimmune bullous disease [1]. Most commonly, this disease is diagnosed between the ages of sixty-six and eighty-three years of age and most frequently in women at a ratio of just over 1:1 to 5:1 [1]. While it can be seen at any age, the incidence of bullous pemphigoid increases from less than thirty people per million people to three hundred and twelve people per million people after age eighty. Furthermore, it is estimated that less than five people in one hundred thousand are affected by this disease [1].

Initial workup for bullous pemphigoid includes a biopsy of the bullae lesion confirming an intact epithelial layer with autoantibodies to the basement membrane. Careful consideration should be used when selecting the site of the skin biopsy. Up to a fifty percent false negative result may occur with specimens sent of the lower extremity. Further workup includes direct and indirect immunofluorescence of the biopsy from the dermal-epidermal junction. Direct immunofluorescence "deposits of IgG occur between 70 and 90% with complement C3 deposition occurring between 90-100%" [2]. Additional workup may include blood work looking for positive autoantibodies, specifically at hemidesmosome antigens BP180 and BP230 [2], and is especially useful in otherwise inconclusive histology. ELISA for antibodies to the BP180 NC16A domain is useful for baseline information in the diagnosis of bullous pemphigoid. Studies have documented sensitivity and specificity of 72 to 90 percent and 90 to 99 percent, respectively. Since IgG BP180 NC16A antibody levels also correlate with disease activity, many clinicians utilize ELISA testing, not only to support the diagnosis of bullous pemphigoid but also as a measure of the response to therapy [3,4].

Current medical therapy includes initial systemic prednisone 1mg/kg/day and DMARDs, specifically methotrexate with or without doxycycline, to prevent a recurrence. While standard oral corticosteroids have traditionally been the mainstay of treatment, topical steroids such as clobetasol propionate cream are just as effective, with potentially fewer side effects and better tolerance among the elderly, with whom this disease generally plagues and can be fatal [5].

Conclusion

Bullous pemphigoid is a disfiguring potentially fatal disorder with a relatively straightforward and often successful treatment course when correctly identified. Nevertheless, clinicians should be mindful and carefully consider this diagnosis even in the setting of nontraditional presentations. Clinicians should further consider additional workups, including a simple antibody blood test BP180 when pathology is inconclusive or clinical suspicion is high. BP180 can also be used as a follow-up dynamic marker of disease activity and response to therapy.

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